

Recurrent Cystic Hygroma

V Ramachandran, P Rajan

Citation

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Abstract

Cystic hygroma is a lymphatic malformation which usually presents in infancy and childhood. When diagnosed by prenatal Ultrasound, it is usually associated with chromosomal anomalies. Although an occasional infant may present with respiratory distress, the major problem is the cosmetic disfigurement they produce. Recurrence following surgical excision is not rare. Here, we present one such case.

CASE REPORT

The patient is a seventeen year old girl who was referred to us with the complaint of a swelling in the right side of her neck for the past six months. She gave a history of previous surgery six years back. After temporary regression, it gradually increased to its present size. On examination, scar of previous surgery was present. There is a diffuse swelling 20 x 15 cms, occupying the posterior triangle of the neck extending to the submandibular and parotid regions, which was smooth, fluctuant, non transilluminant, adherent to the skin and deep to the sternocleidomastoid.

Figure 1

Figure 1: Swelling in neck

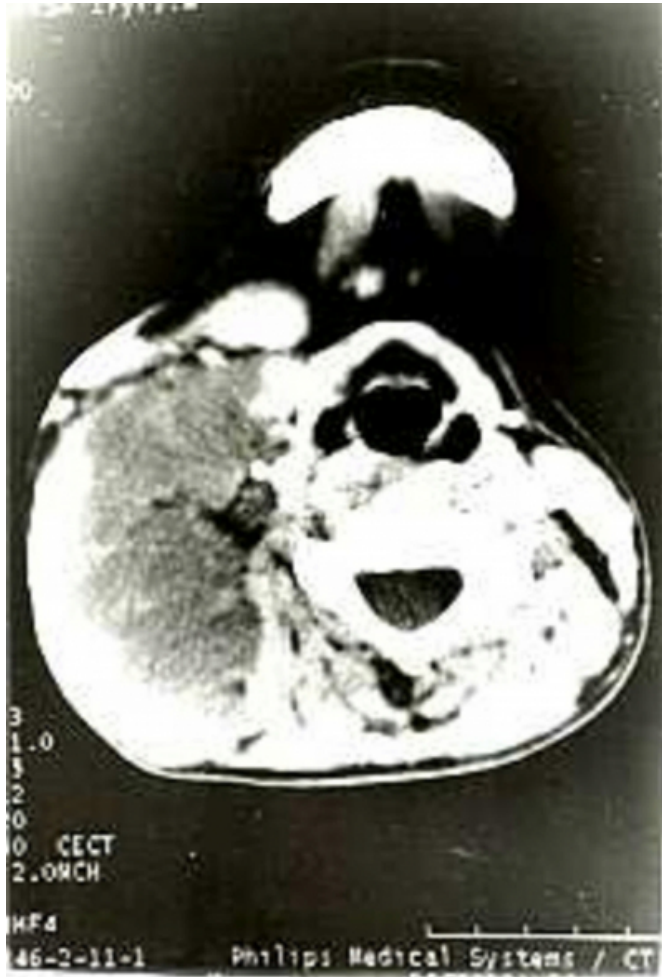


A provisional diagnosis of cystic hygroma was made and a CT – Scan obtained to exclude mediastinal extension.

CT – Scan : Large ,non enhancing,fluid density cystic mass occupying the entire posterior triangle,extending down to just beneath the right clavicle. No extension of the lesion into the mediastinum or axilla. Consistent with recurrent cervical cystic hygroma. (Fig.2)

Figure 2

Figure 2: CT Scan neck



After pre-operative work-up, she was taken up for surgery (Fig.3)

Figure 3



Incision : Transverse cervical

PERIOPERATIVE FINDINGS

A large, multiloculated ,cystic lesion containing haemorrhagic fluid was identified deep to the right sternocleidomastoid extending up to the parotid region superiorly and the right clavicle inferiorly. Excision was done in entire after

dividing the muscle. Haemostasis attained. Suction drain placed after suturing the muscle. (Figs.4 to 7)

Figure 4

Figure 4: Intraoperative view



Figure 5

Figure 5: Intraoperative view



Figure 6

Figure 6: Intraoperative view



Figure 7

Figure 7: Intraoperative view



Her post-operative period was uneventful and she was discharged on the 10th postoperative day.

DISCUSSION

Lymphatic malformations were first described by Redenbacher (1928).Cystic hygroma is one such malformation.

AETIOLOGY

Two theories have been proposed.

- Mc Clure's & Huntington's theory- lymphatic system develops from mesenchymal clefts in venous plexus reticulum and spreads centripetally towards the jugular sac. Lymphatic malformations arise from sequestration or congenital blockage of the primitive lymphatic anlage.
- Sabin's theory- Lymphatic malformations arise from endothelial fibrillar membranes which sprout from the walls of the jugular sac, penetrate surrounding tissue, canalize and produce more cysts.

PATHOLOGY

Lymphatic malformations consist of an aggregation of cysts lined by a single layer of flattened endothelium, filled with lymph, fetal fat and cholesterol crystals.

Classification: Landing & Farber's (1956)

- Lymphangioma simplex-composed of thin walled lymphatic channels.
- Cystic lymphangioma-composed of endothelium lined cysts of varying sizes. e.g : cystic hygroma.

- Cavernous lymphangioma-composed of dilated lymphatics with increased fibrous tissue.

When diagnosed by prenatal ultrasound, lymphatic malformations are frequently associated with chromosomal anomalies. Fetuses with septated cystic hygromas are more commonly associated with Turner's syndrome, more likely to develop hydrops and hence have a poorer prognosis whereas fetuses with non septated cystic hygromas are usually associated with Down's syndrome and have a better prognosis. Other associations include congenital glaucoma, Klippel-Trenaunay

syndrome, lymphangiogenic macroglossia and diaphragmatic hernia (Freyn's syndrome). 60 % occur at birth and 90 % by 2 years. Equal sex incidence exists except for inguinal hygromas which are five times more common in males.

Usual sites include neck, cheek, axilla, groin, mediastinum and the retroperitoneum. The majority of lymphatic malformations in the neck occur in the posterior triangle. Those in the anterior triangle are often associated with intraoral lymphangioma and are the ones likely to produce airway compromise. Mediastinal extension is noted in only 10 % of cases. These swellings are softly cystic, partially compressible (as they are multiloculated) and brilliantly translucent (unless intracystic haemorrhage has occurred).

DIAGNOSIS

Diagnosis is usually made clinically and investigations like chest X-ray, CT-scan and MRI are required only to determine the extent of involvement. The major differential diagnosis is branchial cyst.

Treatment is generally recommended because of the risks of

- Spontaneous infection
- Substantial disfigurement due to progressive growth
- Sudden increase in size with spontaneous haemorrhage
- Airway compromise and dysphagia
- Brachial plexus compression with pain and paraesthesia

TREATMENT OPTIONS

The recurrence rate following surgical extirpation is nearly 10 % and is noted more with those in extra parotid and suprahyoid locations. Most of the recurrences manifest within the first year and are due to residual cysts which grow with the patient. Other complications of surgery include cranial nerve palsies, cosmetic defects, dysphagia and airway compromise.

CORRESPONDENCE TO

Dr. Vijay.R, 19/1931, Bhajanakovil Lane Chalappuram, Calicut Kerala State India Ph: 0091 0495 303205 E - mail: drvijayr1@yahoo.com

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Author Information

Vijay Ramachandran, M.B.B.S.,M.S(Gen.Surgery)-first year

Post graduate student, Department of General Surgery,Calicut Medical College, General Surgery, Calicut Surgical Club

Puthiedath Rajan, M.B.B.S,M.S,F.R.C.S

Associate Professor, Department of General Surgery,Calicut Medical College, General Surgery, Calicut Surgical Club